Major Trigeminal Neuralgia

An Analysis of Two Hundred and Forty-five Cases

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MAJOR trigeminal neuralgia has been a well recognized clinical entity since Fothergill's classical description in 1776.[3] Early in the Nineteenth Century Bell,[1] in England, and Magendie,[10] in France, showed that the fifth cranial nerve rather than the seventh supplied sensation to the face. After this time the treatment of the disease improved rapidly, and it has been more or less standardized since Spiller and Frazier[13] described the procedure used by most neurological surgeons at present. Interestingly, this is one of the few diseases the treatment of which has been known for many years, but the etiology of which remains obscure in spite of considerable investigative work.

A series of 245 cases in our hospital has been reviewed. The disease was found to occur predominantly in the older age groups, the highest incidence being in the decade from 50 to 60 years of age (Figure 1). There were 159 females and 86 males, a ratio of nearly two to one (Figure 2). The right side was involved in 179 cases while the left was involved in only 75; in nine cases (3.6 per cent) there was bilateral involvement (Figure 3). The average duration of symptoms before the patients were seen at this clinic was 7.1 years for females and 7.2 years for males. The longest duration of symptoms was recorded in the case of a woman who had had intermittent episodes of severe pain for 35 years. The onset of pain was approximately equally divided between the second and third divisions of the nerve, onset in the first division being quite rare. The pain often spread to involve adjacent divisions. The second division was involved in 201 cases, the third in 180 instances, and the first in 66 cases (Figure 4). These figures denote only the total number of times pain occurred in the divisions and do not indicate in which division the pain originated.

The description of the pain was strikingly similar in each case. There was a sudden onset of jabbing, cutting, burning, lightning-like or electric-shock-like pain which radiated over one or more divisions of the trigeminal nerve. The radiation was to the peripheral distribution and never crossed the midline. Each individual shock was of short duration, but there might be many shocks in rapid succession. The pain stopped as suddenly as it began. In describing the pain the patients used the same general terms, in which there was a preponderance of adjectives connoting the thermal quality, the cutting sensation, and the suddenness of onset and cessation. As Frazier and Russell[5] have pointed out, the most commonly used adjectives were shooting, sharp, paroxysmal, burning, lightning-like, stabbing, lancinating, and tearing.

Although the first indication of the disease was usually a sudden, typical lancinating pain, there were several patients who noted a peculiar prickling, burning, throbbing sensation for days or weeks prior to the first sharp pain. In one instance a woman had this prickling, throbbing sensation in the upper lip and jaw for five years before the actual onset of paroxysmal pain.

Characteristically there were intervals of complete freedom from pain between the paroxysms, and complete remissions of pain for months or years. With

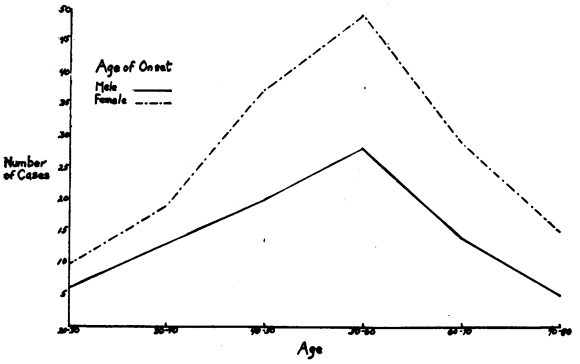

Figure 1

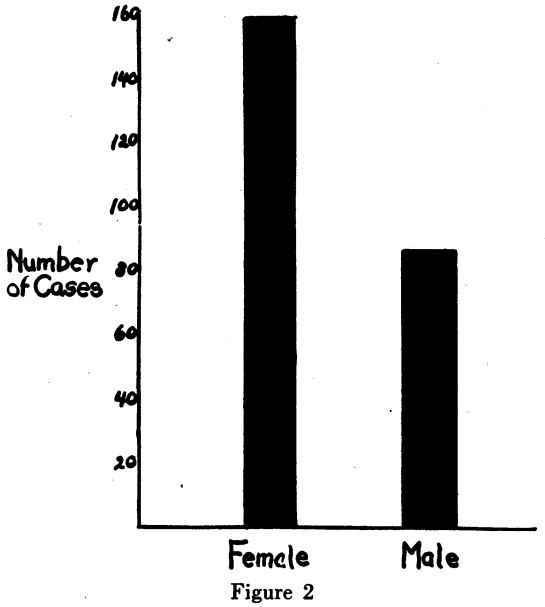

Figure 2

Read before the section on Neuropsychiatry at the 76th Annual Session of the California Medical Association in Los Angeles, April 30-May 3, 1947.

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each remission the patient hoped he would have no more pain, but lived in constant fear of a recurrence.

Usually, but not always, there were definite trigger areas, or small areas which if stimulated would precipitate an attack. These were most commonly on the lips, particularly at the angle of the mouth, the gums, the nasolabial fold, the ala of the nose, and the chin close to the mental foramen. During a severe attack stimulation over any area of the face on the side affected might set off a paroxysm. The trigger area was not always supplied by the same nerve distribution as the affected division. In one instance stimulation of the nasolabial fold precipitated pain in the first division.

The patients were able to describe accurately certain movements or actions which would cause pain. These were most commonly laughing, talking, eating, drinking cold or (sometimes) hot liquids, brushing the teeth, washing the face, or shaving. Many patients would protect their faces on going outdoors, for cold air or wind might give pain. They also learned to shield themselves from these stimuli and to keep the involved side of the face immobile, talking out of the opposite corner of the mouth. They resented examination or any attempt of the examiner to touch the face.

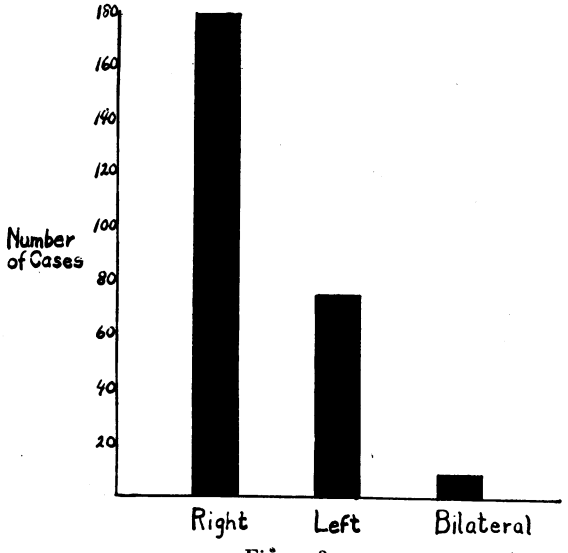

Figure 3

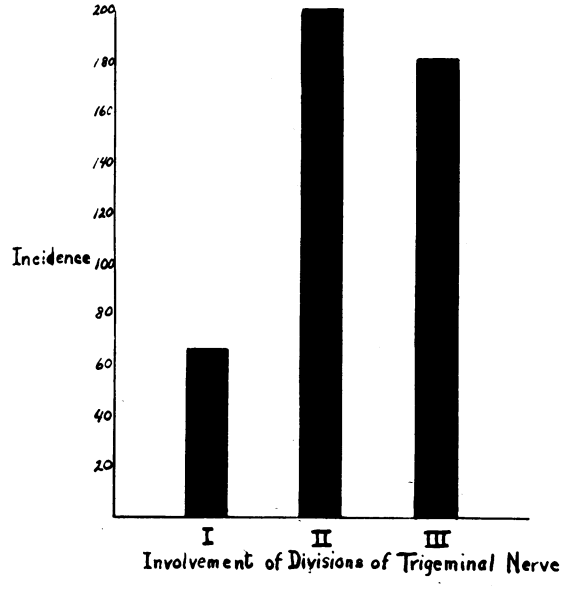

Figure 4

The radiation of the pain varied not only with the division or divisions involved but also in individuals. Often the pain seemed to strike anterior to the ear with explosive violence and then radiate into the peripheral distribution along the upper or lower jaw, into the lips, beneath or above the eye. Radiation into the tongue was not uncommon. In other patients the pain might have its origin in the jaw or gums and radiate upward toward the ear or temple. Many of these people firmly believed that the teeth were the cause of the pain and insisted on extractions. In this series 69 patients (28.1 per cent) had had extractions, usually multiple, in the hope of relieving their pain.

Although the characteristic pain in trigeminal neuralgia is sudden in onset, with cessation and complete freedom from pain between attacks, 44 patients (17.4 per cent) noted residual soreness following an attack, or pain between attacks. Of these, 24 had only residual soreness, aching, or burning sensation, which lasted from minutes to hours after the typical paroxysmal attack of pain. In this group sensory root sections were done in 21 and alcohol injections alone in three cases. All obtained relief. There were postoperative paresthesias in two cases. The remaining 20 of the 44 patients had continuous aching pain between attacks with the paroxysmal lancinating pain superimposed. Sensory root sections were done in 15 cases and alcohol injections alone in five cases. All except one patient obtained complete relief. In this case several alcohol injections were attempted but all were unsuccessful, giving no anesthesia and no relief from pain. A sensory root section was done and this relieved the lancinating pain entirely but did not affect the dull, constant aching pain. There were two cases of mild postoperative paresthesias in this group also.

In 15 cases (6.1 per cent) there were definite manifestations of sympathetic involvement during the attack of pain. In these cases there was flushing of the face, lacrimation, injection of the conjunctiva, or increased salivation. In one case there was marked blanching of the skin over the area of pain in the distribution of the second and third divisions of the trigeminal nerve. All these patients obtained relief from either sensory root section or alcohol injection. There were two cases with mild postoperative paresthesias.

In most instances there was no evidence of predisposing factors. The pain struck suddenly for no apparent reason. However, in 25 cases (10.2 per cent) the patients related the onset of their pain to some definite circumstance. In 19 of these the pain followed a tooth extraction or

other dental work. None of these patients had consulted a dentist because of pain. Typical paroxysmal pain followed the dental work within a period of three hours to two weeks, and the pain centered in the area in which the dental work had been done. In four cases the pain followed sinus infections. In one case the pain followed a blow to the jaw and laceration of the lip, and in another the pain started during a course of x-ray therapy for a basal cell carcinoma of the nose. In these last six cases the typical paroxysmal pain centered in the region which had been previously involved by infection or trauma. All were completely relieved by sensory root section or alcohol injection.

Treatment in this series was primarily surgical. Alcohol injections were done in 175 cases. Many of these patients had multiple injections. The longest period of relief from an alcohol injection was seven years. The average duration of relief was from six to fourteen months. Neurectomies were done in 50 patients. These were mainly supra-orbital, infra-orbital, and inferior alveolar neurectomies, and the period of relief averaged approximately the same as with alcohol injection. Sensory root sections were done in 179 patients. In most of these either alcohol injection or neurectomy had been performed previously. There were three deaths following root section by the subtemporal approach, an operative mortality of 1.6 per cent. Postoperative paresthesias occurred in 27 cases (15 per cent). These consisted of itching, crawling or burning sensations over the anesthetic areas. Some had shooting pains somewhat similar to the original neuralgia but of less severity. The majority of these paresthesias were mild, but in a few instances they were troublesome and persistent. In 32 cases (17.8 per cent) a temporary facial palsy appeared postoperatively, which, however, cleared completely in all instances. The incidence of keratitis in those cases with anesthesia over the first division was low, and available records show only five cases (2.9 per cent). None were severe. Where partial root section was performed pain later appeared in six cases in the undivided portion and required reoperation. In one case of major trigeminal neuralgia involving only the second and third divisions, a differential section was performed, sparing the first division. Seven years later typical pain appeared in the first division and was relieved by complete section of the sensory root. One case with typical shooting pain, but with overflow of the pain into the neck, had no relief from root section. This patient probably had an atypical neuralgia the nature of which might have been shown by alcohol injection preoperatively. Grant [8] had in his series seven similar cases in which he believed alcohol injection would have established the diagnosis and prevented an unsatisfactory operative result.

Trichlorethylene was used in 48 patients in which there was adequate recording of the results. Of these, 25 received some benefit, while 23 could see no change in the character or severity of the pain. In those patients who obtained relief, the usual comment was that it "took the edge off the pain." In some it seemed to hasten a remission, while in a few there was immediate relief. It was noted, however, that often the inhalations would give relief for three to four months and then have no further beneficial effect.

DIAGNOSIS

In the typical case of major trigeminal neuralgia the diagnosis may be made from the history alone. It is confirmed by the absence of neurological changes on examination. As Frazier [4] has pointed out, a diagnosis of trigeminal neuralgia is not justified when there is an associated area of anesthesia or hyperesthesia in the trigeminal zone, when the pain is continuous and not paroxysmal, when there are not periods of complete freedom from pain in the early stages, or when the location of the pain does not correspond to the anatomic zones.

Cushing [2] listed five types of facial neuralgia which might be mistaken for major trigeminal neuralgia. He included sphenopalatine, postherpetic, and geniculate neuralgias, as well as convulsive tic and neuralgias due to tumors. In postherpetic neuralgia there is frequently a sharp shooting pain identical with that in trigeminal neuralgia, but usually there is also a constant burning sensation over the area involved. The history of herpetic lesions and the presence of scarring suggest the diagnosis. The pain in geniculate neuralgia may be differentiated by its location in the ear. Tumors in the cerebello-pontine angle and in or adjacent to the gasserian ganglion may likewise give identical pain (i.e., symptomatic tic) in the early stages. The presence of anesthesia is diagnostic. Sluder [12] has noted that the pain of sphenopalatine neuralgia may occur with stabbing sharpness as in a tic, but with spread of pain over the occiput, neck, and shoulder, and often into the arm. In convulsive tic the paroxysmal pain is associated with the muscular spasm and does not precede it as is the case in a true tic where grimacing may follow the onset of pain.

Many other conditions and their differentiation should be considered. Glossopharyngeal neuralgia causes shooting pain identical with that of trigeminal neuralgia but with its distribution in the pharynx and base of the tongue. Diseases affecting the central nervous system, such as multiple sclerosis, syringomyelia or syringobulbia, and tabes dorsalis, may produce similar pain,[15] but in our experience such cases are extremely rare. Aneurysms involving the sensory root or, more commonly, the first division of the trigeminal nerve usually give rise to a more constant pain associated with sensory or motor changes. Head and facial pain due to vascular disturbances such as the histamine type headache and migraine are distinguishable by the onset and duration of pain. Sinus infections. dental abnormalities and refractive errors ordinarily do not give rise to paroxysmal pain.

In addition, there is a fairly large group of patients whose facial pain is transitional between major trigeminal neuralgia and migraine.[7] It was recognized early that some patients did not get relief from

operative attack on the trigeminal tract in spite of the resulting anesthesia. Frazier and Russell[5] noted certain peculiarities in the type of pain in this group of patients and in 1923 classified such cases as atypical neuralgias. Glaser[6] described the complete syndrome in 1928. The pain is a peculiar, deep-seated ache not referred to the periphery, and is constant and not paroxysmal or intermittent. The syndrome is characterized by attacks of greater or lesser severity at varying intervals. Remissions are rare. It is far more common in females than in males and predominates in the younger age groups. In 50 per cent of the cases there are sympathetic phenomena. The pain does not confine itself to the distribution of any of the cranial nerves, but may involve the scalp as well as the neck and shoulder. In spite of this fairly constant and typical picture, there are instances where the atypical neuralgia may so closely simulate major trigeminal neuralgia that differentiation can be made only by the use of alcohol injections to block the divisions involved. The necessity for accurate diagnosis must be stressed, for it has been thoroughly proved that in atypical neuralgia, as in postherpetic neuralgia, section of the trigeminal nerve not only fails to relieve the pain, but frequently makes the complaints worse.

COMMENTS

Of interest in this series is the group of 44 patients who had residual soreness following paroxysms of pain and continuous pain between paroxysms. Although each had the typical lancinating pain of trigeminal neuralgia, each also had the deep aching pain characteristic of the atypical neuralgias. Since all except one of these patients were completely relieved by alcohol injection or root section, one must conclude that 43 of them had true trigeminal neuralgia.

Similarly, the 15 patients that showed sympathetic phenomena during the paroxysm of pain were all relieved by alcohol injection or root section. Glaser[7] states that sympathetic manifestations occur in 50 per cent of the atypical neuralgias, but are not found in true trigeminal neuralgia. This has not been our experience. In our group the pain was relieved and one must conclude that it was due to major trigeminal neuralgia. Postoperative paresthesias were not more common in these groups than in others with the typical tic.

These two groups serve to emphasize the fact that the clinical picture of trigeminal neuralgia is not always clear-cut and may have features suggestive of atypical neuralgia. It is of particular importance in these cases to make an accurate diagnosis by the use of alcohol injection before performing root sections. If pain is relieved at once when the area is made anesthetic we can be certain of the diagnosis and that section of the root will give lasting relief.

Harris,[9] in his survey of 1,433 cases, states that peripheral sepsis, from such origins as dental caries, pyorrhea, antral abscess and sinusitis, is the prime cause of the disease. No definite relationship of the incidence of trigeminal neuralgia to infection has been shown in other large series,[11] nor has there been any definite relationship to infection in this series. However, the onset of typical trigeminal neuralgia in 19 cases immediately following tooth extraction or dental work seems too frequent an occurrence to be mere coincidence. It would appear that some change had occurred at the peripheral nerve endings which played a part in causing the disease.

The use of trichlorethylene in the treatment of trigeminal neuralgia seemed to be of value in this series, although others[14] have felt the drug was useless in all instances. It proved worthwhile in treating several patients 80 years of age and over, when their condition was such that the operative risk was considered too great and alcohol injections had been unsuccessful. In a few cases the relief was immediate and complete. Whether this was due to regular remissions or not cannot be determined.

Alcohol injections serve two main purposes. If the pain is completely relieved by injection, the diagnosis is established and sensory root section can be carried out with confidence that the pain will be permanently relieved. The anesthesia produced by the injection also serves to accustom the patient to the permanent anesthesia produced by root section. Some patients who were poor operative risks have been kept in comfort by repeated alcohol injections over a period of years.

It is felt that the procedure of choice is a differential section of the sensory root wherever possible with sparing of the motor root. The operative mortality of 1.6 per cent is not unreasonable when one considers that the majority of operations are done on patients between 60 and 75 years of age who often are in a poor state of nutrition because of their inability to eat during their attacks of pain.

DISCUSSION BY NATHAN CROSBY NORCROSS, M.D., OAKLAND

The problem of trigeminal neuralgia has always been a fascinating one to me, more so, perhaps, since the first operation I performed was No. 1002 in Charles Frazier's old operating room. I feel that today we are well equipped to deal rationally and successfully with this disorder. I think the authors of the paper have covered the field thoroughly and with great insight. There are a few comments that I would like to add. The pain of true, major, trigeminal neuralgia is characterized by three things. It is *paroxysmal*; it is *peripheral*; and it is *parallel*—that is, parallel to the anatomical course of the branches of the trigeminal nerve. If these three features are kept in mind, one is unlikely to make a mistake in the diagnosis. As the authors have noted, however, there is an occasional patient apparently having true tic who does not obtain relief from section of the sensory root of the nerve. Even though this percentage is small and has been small in other series of cases, I feel that it is enough to make us want to carry out an alcohol injection before operation is recommended. The alcohol injection is a temporary procedure. The patient will recover from its effects. On the other hand, the operation is permanent and no restoration of function will take place in the nerve involved.

There are several other factors in the use of alcohol injection which I, with the authors, feel make it warranted in practically all cases of this sort. First is the old, debilitated

patient who comes in suffering from severe malnutrition. This is brought on by the intense pain provoked by any attempt to eat or drink. This patient is in no condition for operation and should be given temporary relief by the simplest method possible in order that he can be built up and got into condition for later operation. Second: Occasionally the differential diagnosis between true tic and atypical facial neuralgia or a combination of neuralgias is extremely difficult and, in these cases, alcohol injection is the diagnostic agent of choice. Third is the patient who will have severe paresthesias following root section. These patients cannot be segregated from the others until an alcohol injection or the operation has been performed. To be sure, they constitute a small percentage, perhaps between 3 and 5 per cent. They state that the paresthesias are almost or fully as severe as the pain they had prior to operation. I feel that the classical, subtemporal approach of Frazier is probably not the most desirable procedure in this small group, but that we should consider trigeminal tractotomy which leaves sensation in the face almost normal. So far as we know at present, this procedure offers a better chance of avoiding severe, incapacitating, postoperative paresthesia.

Trichlorethylene has been found to be a helpful adjunct in the early treatment of this condition in some patients and I, empirically, add to that large doses of B-1. With trichlorethylene I have been able to bring about what appears to be spontaneous remissions from tic for fairly long periods of time. This brings up a point that I feel about quite strongly. In the more elderly group of patients I do not feel we are warranted in rushing ahead to recommend major surgery. To be sure, the mortality rate of this procedure is only between 1 and 2 per cent and this has been essentially true for all of the various reports of large groups that have appeared in the literature. One must remember, however, that in many of these elderly patients some of the less radical methods will relieve the pain for the duration of their lives, without their having to be subjected to a major surgical procedure.

In regard to the possible etiology of tic about which, as the authors have said, nothing conclusive is known, one hint has been brought out by Lewy and Grant, from their pneumoencephalographic and histological studies of patients with major trigeminal neuralgia. Histologically, in six cases it was demonstrated that there were degenerative changes of the thalamic nuclei and of the corona radiata, bringing about some degree of interruption of the thalamocortical fibers. The surprising thing about their findings was that in five cases out of six these lesions were found only on the same side as the tic had been. Encephalograms had also been performed at the University of Pennsylvania during that time and several of these demonstrated atrophic changes deep in the ipsilateral hemisphere, apparently caused by wasting of the thalamus. Also, attention was called to the fact that of one group of 50 cases investigated thoroughly, 30 showed definite evidence of arteriosclerosis and renal dysfunction.

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